ORIGINAL ARTICLE

Changing Pattern of Ectopic Pancreas: 22 Years of Experience in a Medical Center

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Background/Purpose: Ectopic pancreas is usually a silent gastrointestinal malformation. It may become clinically evident when complicated by chronic inflammation or by growth. More ileal ectopic pancreas has been found in recent years in our hospital. We report the clinical manifestation of ectopic pancreas over the past 22 years.

Methods: We reviewed the medical records of patients seen between May 1984 and December 2005 at Mackay Memorial Hospital, with a diagnosis of ectopic pancreas, and extracted clinical and histopathology data from the records.

Results: A total of 39 patients (18 male, 21 female; mean age, 46 years) were diagnosed with ectopic pancreas. Most patients were aged between 30 and 50 years. Only 15 (38%) had symptoms suggestive of ectopic pancreas. These included abdominal pain ($n=9$), upper gastrointestinal bleeding ($n=5$), and abdominal distension ($n=2$). The diagnosis in the other 24 patients was made incidentally, usually during surgery for other conditions. While lesions in the stomach were more likely to be diagnosed because of symptoms (12 of 13), lesions in the small bowel were almost always diagnosed incidentally. Only one of eight in the duodenum, one of 10 in the jejunum, and one of eight in the ileum, were isolated findings. One case of ectopic pancreas was detected by capsule endoscopy.

Conclusion: Ectopic pancreas can be found in various parts of the gastrointestinal tract. The high proportion of ileal ectopic pancreas is unexpected and needs further study. [*J Formos Med Assoc* 2008;107(12): 932–936]

Key Words: capsule endoscopy, double-balloon enteroscopy, ectopic pancreas, gastrointestinal tract, small intestine

Ectopic pancreas (also called pancreatic heterotopia, aberrant pancreas, heterotopic pancreas, pancreatic rest, or accessory pancreas) is tissue histologically similar to normal pancreatic tissue but found elsewhere than its usual location, which has no anatomic or vascular connection with the pancreas itself.[1–3] Schiltz reported the first example in 1727, and Klob described the histology of this condition in 1859.[4,5] It is often an incidental finding at various sites in the gastrointestinal (GI) tract. The incidence at autopsy ranges from 0.5% to 13%, with 70–90% of the lesions found in the stomach, duodenum or jejunum.[6–8] In recent years, however, we have found more cases of ileal ectopic pancreas. The purpose of this study was to retrospectively review our experience with

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ectopic pancreas, with particular attention to the location and methods of diagnosis.

Patients and Methods

We retrieved the records of all patients seen between May 1984 and December 2005 at Mackay Memorial Hospital, Taipei, who had a diagnosis of ectopic pancreas. The confirmative diagnosis was made by a pathologist. Each pathological slide that had been stained with hematoxylin and eosin was reviewed to determine whether the components of pancreatic tissue, including acini, ducts and islets of Langerhans were present. We reassessed the pathologic slides for the study with another pathologist. During this period, there were 15 cases of gastric lesions seen endoscopically that resembled ectopic pancreas. However, no tissue diagnosis was made, and these patients were not included. Patient data extracted from the clinical records included age, gender, symptoms, diagnostic methods, tentative preoperative diagnosis, treatment, and follow-up. If ectopic pancreas was confirmed pathologically and the clinical manifestations were relieved by its removal, the lesion was classified as being in the isolated symptomatic group. In those cases associated with other pathologic processes, ectopic pancreas was classified in the incidental group. Follow-up data were completed either according to the latest visit at the outpatient department or by telephone. Statistical analysis of continuous variables was performed using a two-tailed Student's *t* test.

Results

Over the 22-year study period, 39 patients (18 male, 21 female) were diagnosed with ectopic pancreas. The incidence increased over that time, particularly in the final 5 years of the study period (Figure 1). Most strikingly, there were 15 ectopic lesions found from 2001 to 2005, of which six were in the ileum. This contrasted with only two ileal lesions identified in the preceding 15 years. The patients' mean age was 46 years (range, 3–79 years), with 14 patients aged 30–50 years (Figure 2). While the mean age of men was slightly lower than that of women, the difference was not significant (mean, 43 *vs.* 49 years, $p = 0.32$, χ^2 test). Only 15 patients had symptoms that were attributed to ectopic pancreas, including abdominal pain ($n = 9$), upper GI bleeding ($n = 5$), abdominal distension ($n = 2$), and weight loss ($n = 1$). These patients were classified as having isolated symptomatic lesions. In the remaining 24 patients, ectopic pancreas was found during

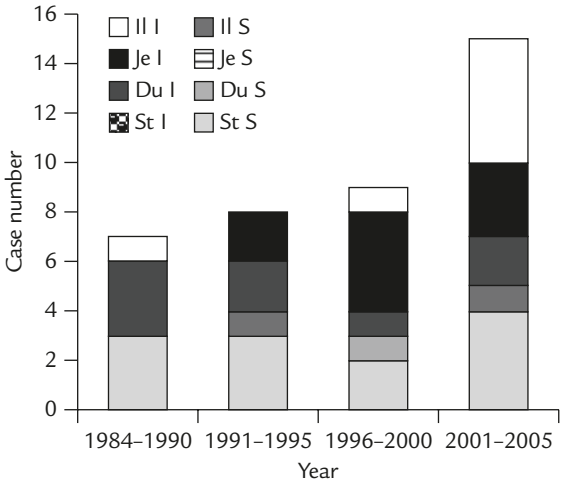

Figure 1. Number of cases of ectopic pancreas by 5-year periods and site. Il = ileum; Je = jejunum; Du = duodenum; St = stomach; I = incidental group; S = symptomatic group.

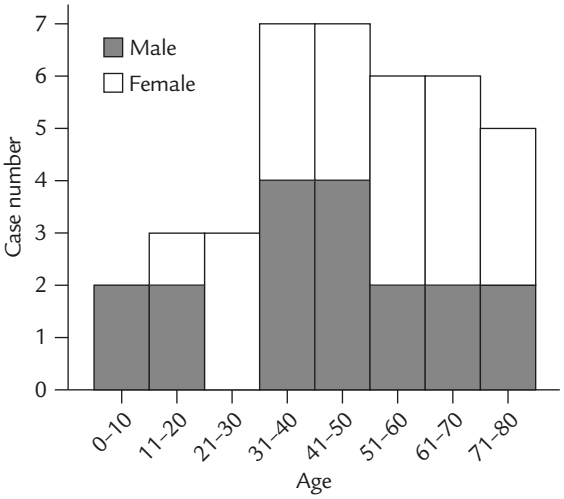

Figure 2. Age and gender distribution of patients with ectopic pancreas.

surgery for other conditions (Table 1). In these cases, a palpable mass in the GI tract was incidentally noted at operation and removed.

A third of the lesions were located in the stomach, with the remaining two thirds found in the small bowel (Table 2). Three of eight ileal lesions were found in the Meckel diverticula. Isolated lesions were significantly more likely to be found in the stomach than were those discovered incidentally (12/15, 80% *vs.* 1/24, 4%, $p < 0.05$). Lesions were found by gastroduodenoscopy in 13 patients, 11 in the stomach and two in the duodenum. Of these, however, only four had a correct pathologic diagnosis made on an endoscopic biopsy specimen alone, and all of them received surgical intervention later. In the remainder, diagnosis was made only after surgery. Barium upper GI series was performed in 14 patients, three of whom were found to have submucosal tumor, two in the stomach and one a 5.5-cm lesion in the jejunum. In the final 5-year period, a patient with GI bleeding whose source was obscure was seen to have a polyp in the ileum upon capsule endoscopy, which subsequently proved to be ectopic pancreas.

The preoperative diagnosis in 10 (26%) patients was submucosal tumor, based on gastroduodenoscopy. Subtotal gastrectomy was performed in eight patients, rather than local excision, because malignancy was suspected. Only one of these patients actually had malignant gastric adenocarcinoma, and another was thought to have malignant potential. Surgery for the incidentally discovered lesions, for the most part, involved only segmental resection with primary anastomosis.

Other investigations, such as abdominal ultrasound, computed tomography, and angiography did not disclose the tumors in most patients. The exact size of the lesions was clearly recorded for only 29 patients. The mean diameter was 1.1 cm (range, 0.1–5.5 cm), with 15 lesions < 1 cm. Definitive diagnosis required pathological examination. The lesion types were classified according to modified von Heinrich's classification: mixed type II and type III (acini and ducts) was most common ($n = 9$), followed by type II ($n = 5$) and type I (all pancreatic cell types; $n = 2$). Some patients had malignancy not related to ectopic pancreas, including: gastric adenocarcinoma ($n = 2$), colon cancer ($n = 1$), sigmoid cancer ($n = 1$), rectal cancer ($n = 1$), GI stroma tumor ($n = 1$), myelodysplastic syndrome ($n = 1$), ampulla of Vater cancer

Table 1. Reasons for operation in 24 patients with incidentally discovered ectopic pancreas

Disease	Number
Intussusception	6*
Intestinal obstruction	2†
Cholecystitis	2
Acute appendicitis	2
Ampulla of Vater cancer	2
Gastric cancer	2
Splenic tumor	1
Peritonitis	1
Crohn's disease with fistula	1
Adhesion ileus	1
Chronic peptic ulcer disease with obstruction	1
Colon cancer	1
Sigmoid cancer	1
Rectal cancer	1

**Including one case of gastrointestinal stroma tumor; †including one case of uterine malignant mixed Mullerian tumor.*

Table 2. Location of ectopic pancreas in isolated or incidental finding

Location	Isolated, *n*	Incidental, *n*	Total, *n* (%)
Stomach	12	1	13 (33)
Duodenum	1	7	8 (21)
Jejunum	1	9	10 (25)
Ileum	1	7	8 (21)
Total	15	24	39 (100)

($n=2$), and uterine malignant mixed Mullerian tumor ($n=1$).

Discussion

In this 22-year series, the incidence of ectopic pancreas was usually 7–9 cases every 5 years, until the most recent 5-year period, when 15 patients were diagnosed, three quarters of whom had lesions in the small bowel, including six in the ileum. The incidence of ectopic pancreas in the literature ranges from 0.55% to 13.7% in autopsy series, with most reports being at the lower end of this range.[3–5,8,9] Since it is usually an incidental finding, the prevalence of ectopic pancreas may well be higher than reported figures suggest. The interesting finding in our study was the relative increase in the number of small bowel lesions, particularly in the ileum, toward the end of the study period. In the past, most studies have reported 70–90% of lesions being in the upper gut, including the stomach (25–38.2%), duodenum (17–36.3%), and jejunum (15–21.7%).[3,5,8,10] Uncommonly reported locations have included the ileum, colon, spleen, liver, biliary tract, mesentery and lymph nodes.[3,11,12] The 21% incidence of ileal lesions in our series is thus surprising. These lesions are most often discovered incidentally during radiographic or endoscopic examination of the gut, at surgical exploration, or autopsy.[13,14] Our data lead us to speculate whether the incidence of ileal ectopic pancreas has increased.

In our series, the majority of isolated ectopic pancreas lesions found because of symptoms were in the upper GI tract. The symptoms in our series were similar to those reported by others, including abdominal pain, nausea, vomiting, anemia, weight loss and melena.[1,3,5,13,15] These complaints are so nonspecific that they are of little help in suggesting the correct diagnosis. However, investigation undertaken because of symptoms leads to identification of the lesion. This is obviously easier to accomplish in the upper GI tract, where ectopic pancreas may have a characteristic appearance. When seen on gastroduodenoscopy, it is usually a well-circumscribed, soft, rubbery, yellow, submucosal or intramural mass with central umbilication.[3,5,7,8] Upon barium meal examination, it appears typically as a well-delineated submucosal filling defect, with a characteristic central indentation.[1,3–5] The distal small bowel, on the other hand, is notoriously difficult to visualize in its entirety, therefore it is not surprising that ectopic pancreas in the ileum is found rarely other than incidentally. The one patient in our series in whom this was not true underwent capsule endoscopy to investigate GI bleeding, a case we have reported previously.[3]

Capsule endoscopy, a new technique for visualizing the small bowel, has high sensitivity and specificity for detecting sources of bleeding in patients with obscure GI bleeding and small bowel lesions.[3,16] A German multicenter trial of 56 patients found a better diagnostic yield with capsule endoscopy than with push enteroscopy, enteroclysis, or angiography.[17] The technique provides high-quality images with a negligible rate of adverse events and complications.[3,16,18] This may well become the procedure of choice for evaluation of patients with suspected small bowel lesions. The main disadvantage is that it does not allow for biopsy. Another technique, double balloon enteroscopy, provides high-resolution visualization plus diagnostic and therapeutic procedures in all segments of the small intestine.[19,20] A group of investigators in Hungary has reported a small pilot study using this technique, and identified small bowel lesions in 12 of 22 patients.[21] It remains to be seen whether capsule or double-balloon endoscopy, or perhaps other techniques developed in the future, will lead to increased discovery of ectopic pancreas in the lower small bowel.

In conclusion, diagnosis of ectopic pancreas is extremely difficult to make preoperatively, even in symptomatic cases. Whether incidentally found lesions would have become symptomatic if left in place is a moot point. The recent increase in the incidence of ileal lesions in our series is as yet unexplained. However, we expect that improved methods of visualizing the small bowel may well

lead to the finding of even more cases of ectopic pancreas in the small bowel.

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